UNITED STATES
SECURITIES AND EXCHANGE COMMISSION
WASHINGTON, D.C. 20549

FORM 8-K

CURRENT REPORT
Pursuant to Section 13 or 15(d) of the Securities Exchange Act of 1934

Date of Report (Date of earliest event reported): **January 4, 2024**

Kiniksa Pharmaceuticals, Ltd.
(Exact name of Registrant as Specified in Its Charter)

Bermuda	**001-730430**	**98-1327726**
(State or other jurisdiction of incorporation or organization)	(Commission File Number)	(I.R.S. Employer Identification No.)

Kiniksa Pharmaceuticals, Ltd.
Clarendon House
2 Church Street
Hamilton HM11, Bermuda
(808) 451-3453
(Address, zip code and telephone number, including area code of principal executive offices)

Kiniksa Pharmaceuticals Corp.
100 Hayden Avenue
Lexington, MA, 02421
(781) 431-9100
(Address, zip code and telephone number, including area code of agent for service)

N/A
(Former Name or Former Address, if Changed Since Last Report)

Check the appropriate box below if the Form 8-K filing is intended to simultaneously satisfy the filing obligation of the registrant under any of the following provisions

☐ Written communications pursuant to Rule 425 under the Securities Act (17 CFR 230.425)

☐ Soliciting material pursuant to Rule 14a-12 under the Exchange Act (17 CFR 240.14a-12)

☐ Pre-commencement communications pursuant to Rule 14d-2(b) under the Exchange Act (17 CFR 240.14d-2(b))

☐ Pre-commencement communications pursuant to Rule 13e-4(c) under the Exchange Act (17 CFR 240.13e-4(c))

Securities registered pursuant to Section 12(b) of the Act:

Title of each class	**Trading Symbol(s)**	**Name of each exchange on which registered**
Class A Common Shares $0.000273235 par value	KNSA	The Nasdaq Stock Market LLC (Nasdaq Global Select Market)

Indicate by check mark whether the registrant is an emerging growth company as defined in Rule 405 of the Securities Act of 1933 (§230.405 of this chapter) or Rule 12b-2 of the Securities Exchange Act of 1934 (§240.12b-2 of this chapter).

Emerging growth company ☐

If an emerging growth company, indicate by check mark if the registrant has elected not to use the extended transition period for complying with any new or revised financial accounting standards provided pursuant to Section 13(a) of the Exchange Act. ☐

Item 2.02. Results of Operations and Financial Condition.

On January 4, 2024, Kiniksa Pharmaceuticals, Ltd. (the "Company") issued a press release (the "Press Release") announcing, among other things, that (i) its preliminary year-end 2023 cash, cash equivalents and short-term investments of $206.3 million (unaudited) are expected to fund its current operating plan into at least 2027 and (ii) ARCALYST net revenue was $71.2 million and $233.1 million for the fourth quarter and full year 2023, respectively (unaudited).

The preliminary selected financial results reported by the Company are unaudited, subject to adjustment, and provided as an approximation in advance of the Company's expected announcement of complete financial results in February 2024.

Item 7.01. Regulation FD Disclosure.

In addition to the information contained in Item 2.02, the Press Release also announced top-line data from Cohorts 1, 2 and 3 of the Company's Phase 2 clinical trial of abiprubart (KPL-404) in rheumatoid arthritis. In connection with such announcement, the Company posted an investor presentation (the "Investor Presentation") containing data from the trial to its website at investors.kiniksa.com.

A copy of the Press Release and the Investor Presentation are furnished with this Current Report on Form 8-K as Exhibit 99.1 and Exhibit 99.2, respectively.

The information contained in these Items 2.02 and 7.01 of this Current Report on Form 8-K and Exhibits 99.1 and 99.2 shall not be deemed "filed" for purposes of Section 18 of the Securities Exchange Act of 1934, as amended (the "Exchange Act"), or otherwise subject to the liabilities of that section, nor shall it be deemed incorporated by reference in any filing under the Securities Act of 1933, as amended, or the Exchange Act, regardless of any general incorporation language in such filing and except as expressly provided by specific reference in such filing.

Item 9.01. Financial Statements and Exhibits.

(d) Exhibits

Exhibit No.	Description
99.1	Press Release issued by Kiniksa Pharmaceuticals, Ltd., dated January 4, 2024
99.2	Kiniksa Pharmaceuticals, Ltd. Investor Presentation
104	Cover Page Interactive Data File (embedded within the inline XBRL document)

SIGNATURES

Pursuant to the requirements of the Securities Exchange Act of 1934, the registrant has duly caused this report to be signed on its behalf by the undersigned hereunto duly authorized.

KINIKSA PHARMACEUTICALS, LTD.

Date: January 4, 2024

By: /s/ Madelyn Zeylikman
Madelyn Zeylikman
Senior Vice President, General Counsel and Secretary

Kiniksa Pharmaceuticals Provides Corporate Update

– ARCALYST® (rilonacept) 2023 net product revenue grew ~90% year-over-year to $233.1 million (unaudited) –

– ARCALYST 2024 net product revenue expected to be $360 - $380 million –

– Abiprubart (KPL-404) Phase 2 rheumatoid arthritis trial met the primary efficacy endpoint in Cohort 3 at the weekly dose level –

– Abiprubart Phase 2 rheumatoid arthritis data from Cohort 4 expected in Q2 2024 –

– Cash reserves of $206.3 million (unaudited) expected to fund operations into at least 2027–

HAMILTON, BERMUDA – January 4, 2024 – Kiniksa Pharmaceuticals, Ltd. (Nasdaq: KNSA) (Kiniksa), a commercial-stage biopharmaceutical company with a pipeline of immune-modulating assets designed to target a spectrum of cardiovascular and autoimmune diseases, today provided a corporate update.

"Strong execution to date has laid the foundation for the continued advancement of Kiniksa's portfolio in 2024. ARCALYST 2023 net product revenue grew ~90% year-over-year to $233.1 million, underscoring our robust commercial performance. We believe there is substantial opportunity with ARCALYST in recurrent pericarditis and expect to drive continued revenue growth and collaboration profitability by reaching an increasing number of patients. In fact, at the end of 2023 Kiniksa penetrated approximately 9% into the multiple-recurrence population, compared to approximately 5% at the end of 2022," said Sanj K. Patel, Chairman and Chief Executive Officer of Kiniksa. "Additionally, abiprubart showed clinical effect in the first three cohorts of the Phase 2 trial in rheumatoid arthritis. Despite a high placebo response rate, the 5 mg/kg weekly dose level in Cohort 3 achieved statistical significance, but the 5 mg/kg biweekly dose level did not. We look forward to evaluating results from Cohort 4, and we will use the totality of the data to determine next steps for the program. Importantly, our strong financial position provides optionality to continue to invest across our business, including ARCALYST commercialization as well as both pipeline and business development."

Portfolio Execution

ARCALYST (IL-1α and IL-1β cytokine trap)

- ARCALYST net product revenue was $71.2 million and $233.1 million for the fourth quarter and full year 2023, respectively (unaudited).
- Since launch in April 2021, more than 1,700 prescribers have written ARCALYST prescriptions for recurrent pericarditis.

- As of the end of the fourth quarter of 2023, average total duration of ARCALYST therapy in recurrent pericarditis increased to approximately 23 months.
- As of the end of the fourth quarter of 2023, approximately 9% of the target 14,000 multiple-recurrence patients were actively on ARCALYST treatment.
- Kiniksa increased the size of its salesforce to approximately 85 representatives by the end of 2023 to help drive further physician adoption and patient enrollments in 2024.
- Kiniksa expects 2024 ARCALYST net product revenue of between $360 million and $380 million.

Abiprubart (anti-CD40 monoclonal antibody inhibitor of CD40-CD154 interaction)

- Kiniksa today announced that the Phase 2 clinical trial of abiprubart in rheumatoid arthritis met its primary efficacy endpoint, change from baseline in Disease Activity Score of 28 Joints Using C-reactive Protein (DAS28-CRP) versus placebo.
    - In Cohorts 1 and 2 (pharmacokinetic (PK)-lead in), multiple doses of abiprubart were well-tolerated and enabled the proof-of-concept portion of the study. Although these cohorts were not powered for DAS28-CRP (Secondary Efficacy Endpoint), the following results were observed:
        - In Cohort 1, in the abiprubart 2 mg/kg subcutaneous (SC) biweekly dosing group (n=6), the mean change from baseline in DAS28-CRP at Week 12 was -3.16 points compared to -1.09 points in pooled placebo recipients (n=4), (Mean Difference = -2.07, p=0.0312).
        - In Cohort 2, in the abiprubart 5 mg/kg SC biweekly dosing group (n=6), the mean change from baseline in DAS28-CRP at Week 12 was -3.44 points compared to pooled placebo recipients (n=4), (Mean Difference = -2.35, p=0.0338).
    - In Cohort 3, in the abiprubart 5 mg/kg SC weekly dosing group (n=27), the Least Squares (LS) mean change [95% confidence interval (CI)] from baseline in DAS28-CRP at Week 12 was -2.21 [-2.62, -1.80] points compared to -1.65 [-2.07, -1.23] points in placebo recipients (n=26), (LS Mean Difference = -0.56, p=0.0487).
    - In Cohort 3, in the abiprubart 5 mg/kg SC biweekly dosing group (n=25), the LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -2.00 [-2.43, -1.58] points compared to -1.65 [-2.07, -1.23] points in placebo recipients (n=26), (LS Mean Difference = -0.35, p=0.2140).
    - Abiprubart significantly reduced Rheumatoid Factor (a clinical marker of disease activity and autoantibody pharmacodynamic marker of CD40 target engagement) by over 40% in both Cohort 3 dose levels.
    - Abiprubart was well-tolerated, with no dose-related adverse experiences observed.
- Kiniksa has now completed enrollment in a fourth cohort (Cohort 4) of the Phase 2 clinical trial of abiprubart in rheumatoid arthritis. Cohort 4 will evaluate a fixed dose level administered as a single subcutaneous injection once monthly. The company expects data from Cohort 4 in the second quarter of 2024.

Mavrilimumab (monoclonal antibody inhibitor targeting GM-CSFRα)

- Kiniksa is now evaluating potential partnership opportunities to advance development of mavrilimumab, which has generated positive data in mid-stage clinical trials across multiple indications.

Corporate Update

- Kiniksa's year-end 2023 cash, cash equivalents, and short-term investments of $206.3 million (unaudited) are expected to fund its current operating plan into at least 2027.

42nd Annual J.P. Morgan Healthcare Conference

- Sanj K. Patel, Chief Executive Officer and Chairman of the Board of Kiniksa will provide a corporate presentation at the 42nd Annual J.P. Morgan Healthcare Conference on January 8, 2024, at 1:30 p.m. Pacific Time (4:30 p.m. Eastern Time). A live webcast of Kiniksa's presentation will be accessible through the Investors & Media section of the company's website at www.kiniksa.com. A replay of the webcast will also be available on Kiniksa's website within approximately 48 hours after the event.

About Kiniksa

Kiniksa is a commercial-stage biopharmaceutical company focused on discovering, acquiring, developing, and commercializing therapeutic medicines for patients suffering from debilitating diseases with significant unmet medical need. Kiniksa's immune-modulating assets, ARCALYST, abiprubart, and mavrilimumab, are based on strong biologic rationale or validated mechanisms, target a spectrum of underserved cardiovascular and autoimmune conditions, and offer the potential for differentiation. For more information, please visit www.kiniksa.com.

About ARCALYST

ARCALYST is a weekly, subcutaneously injected recombinant dimeric fusion protein that blocks interleukin-1 alpha (IL-1α) and interleukin-1 beta (IL-1β) signaling. ARCALYST was discovered by Regeneron Pharmaceuticals, Inc. (Regeneron) and is approved by the U.S. Food and Drug Administration (FDA) for recurrent pericarditis, cryopyrin-associated periodic syndromes (CAPS), including Familial Cold Autoinflammatory Syndrome and Muckle-Wells Syndrome, and deficiency of IL-1 receptor antagonist (DIRA). The FDA granted Breakthrough Therapy designation to ARCALYST for the treatment of recurrent pericarditis in 2019 and Orphan Drug exclusivity to ARCALYST in 2021 for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and pediatric patients 12 years and older. The European Commission granted Orphan Drug Designation to ARCALYST for the treatment of idiopathic pericarditis in 2021.

IMPORTANT SAFETY INFORMATION ABOUT ARCALYST

- ARCALYST may affect your immune system and can lower the ability of your immune system to fight infections. Serious infections, including life-threatening infections and death, have happened in patients taking ARCALYST. If you have any signs of an infection, call your doctor right away. Treatment with ARCALYST should be stopped if you get a serious infection. You should not begin treatment with ARCALYST if you have an infection or have infections that keep coming back (chronic infection).
- While taking ARCALYST, do not take other medicines that block interleukin-1, such as Kineret® (anakinra), or medicines that block tumor necrosis factor, such as Enbrel® (etanercept), Humira® (adalimumab), or Remicade® (infliximab), as this may increase your risk of getting a serious infection.
- Talk with your doctor about your vaccine history. Ask your doctor whether you should receive any vaccines before you begin treatment with ARCALYST.
- Medicines that affect the immune system may increase the risk of getting cancer.
- Stop taking ARCALYST and call your doctor or get emergency care right away if you have any symptoms of an allergic reaction.
- Your doctor will do blood tests to check for changes in your blood cholesterol and triglycerides.
- Common side effects include injection-site reactions (which may include pain, redness, swelling, itching, bruising, lumps, inflammation, skin rash, blisters, warmth, and bleeding at the injection site), upper respiratory tract infections, joint and muscle aches, rash, ear infection, sore throat, and runny nose.

For more information about ARCALYST, talk to your doctor and see the Product Information.

About Abiprubart (KPL-404)

Abiprubart (KPL-404) is an investigational humanized monoclonal antibody that binds to CD40 and is designed to inhibit the CD40-CD154 (CD40 ligand) interaction, a key T-cell co-stimulatory signal critical for B-cell maturation and immunoglobulin class switching and Type 1 immune responses. Kiniksa believes disrupting the CD40-CD154 co-stimulatory interaction is an attractive approach to addressing multiple autoimmune disease pathologies.

About the Phase 2 Clinical Trial of Abiprubart in Rheumatoid Arthritis

The ongoing Phase 2 rheumatoid arthritis trial is a randomized, double-blind, placebo-controlled trial designed to evaluate pharmacokinetics, safety, and efficacy of chronic subcutaneous administration of abiprubart and to provide optionality to evaluate abiprubart across a range of autoimmune diseases. This trial enrolled patients with active rheumatoid arthritis who had an inadequate response or were intolerant to a Janus kinase inhibitor (JAKi) or at least one biologic disease-modifying anti-rheumatic drug (bDMARD).

The multiple ascending-dose PK lead-in portion randomized 8 patients each in a 3:1 ratio to receive abiprubart 2 mg/kg or placebo (Cohort 1) or 5 mg/kg or placebo (Cohort 2), administered subcutaneously biweekly over a period of 12 weeks. The primary objective of this part of the trial was to evaluate pharmacokinetics, safety, and tolerability over 12 weeks. The secondary efficacy endpoint was change from baseline in DAS28-CRP versus placebo.

The first part of the proof-of-concept portion of the trial (Cohort 3) randomized 78 patients in a 1:1:1 ratio to receive abiprubart 5 mg/kg SC weekly, abiprubart 5 mg/kg SC biweekly, or placebo over a period of 12 weeks. The final part of the proof-of-concept portion of the trial (Cohort 4) randomized 51 patients in a 3:2 ratio to receive a fixed 600 mg loading dose on Day 1 followed by 400 mg SC every four weeks or placebo over a period of 12 weeks. The primary efficacy endpoint of the proof-of-concept portion of the trial is change from baseline in DAS28-CRP versus placebo.

About Mavrilimumab

Mavrilimumab is an investigational fully human monoclonal antibody that blocks activity of GM-CSF by specifically binding to the alpha subunit of the GM-CSF receptor (GM-CSFRα). Phase 2 clinical trials of mavrilimumab in rheumatoid arthritis and giant cell arteritis achieved their primary and secondary endpoints with statistical significance. Kiniksa is now evaluating potential partnership opportunities for mavrilimumab.

Forward-Looking Statements

This press release contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995. In some cases, you can identify forward looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "target," "project," "contemplate," "believe," "estimate," "predict," "potential" or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. All statements contained in this press release that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation, statements regarding: our expectation that ARCALYST 2024 net product revenue will be between $360 million and $380 million; our plan to report data from Cohort 4 of our Phase 2 clinical trial of abiprubart in rheumatoid arthritis in the second quarter of 2024; our expectation about our cash reserves funding our current operating plan into at least 2027; our expectation that we will drive continued ARCALYST revenue growth and collaboration profitability by reaching an increasing number of patients; our beliefs about the mechanisms of our product candidates and potential impact of their approach, including that using abiprubart to disrupt the CD40-CD154 co-stimulatory interaction is an attractive approach to address multiple autoimmune disease pathologies; and our belief that all of our product candidates offer the potential for differentiation.

These forward-looking statements are based on management's current expectations. These statements are neither promises nor guarantees, but involve known and unknown risks, uncertainties and other important factors that may cause our actual results, performance or achievements to be materially different from any future results, performance or achievements expressed or implied by the forward-looking statements, including without limitation, the following: delays or difficulty in enrollment of patients in, and activation or continuation of sites for, our clinical trials; delays or difficulty in completing our clinical trials as originally designed; potential for changes between final data and any preliminary, interim, top-line or other data from clinical trials; our inability to replicate results from our earlier clinical trials or studies; impact of additional data from us or other companies, including the potential for our data to produce negative, inconclusive or commercially uncompetitive results; potential undesirable side effects caused by our products and product candidates; our inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for applicable regulatory authorities to not accept our filings, delay or deny approval of any of our product candidates or require additional data or trials to support approval; inability to successfully execute on our commercial strategy for ARCALYST; our reliance on third parties as the sole source of supply of the drug substance and drug product used in our products and product candidates; our reliance on Regeneron as the current sole manufacturer of ARCALYST; risks arising from our ongoing technology transfer of ARCALYST drug substance manufacturing; raw material, important ancillary product and drug substance and/or drug product shortages; our reliance on third parties to conduct research, clinical trials, and/or certain regulatory activities for our product candidates; complications in coordinating requirements, regulations and guidelines of regulatory authorities across jurisdictions for our clinical trials; changes in our operating plan, business development strategy or funding requirements; and existing or new competition.

These and other important factors discussed in our filings with the U.S. Securities and Exchange Commission, including under the caption "Risk Factors" contained therein, could cause actual results to differ materially from those indicated by the forward-looking statements made in this press release. Any such forward-looking statements represent management's estimates as of the date of this press release. Except as required by law, we disclaim any intention or obligation to update or revise any forward-looking statements. These forward-looking statements should not be relied upon as representing our views as of any date subsequent to the date of this press release.

Every Second Counts! ®

Kiniksa Investor and Media Contact

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Corporate Presentation

JANUARY 2024

Forward Looking Statements

This presentation (together with any other statements or information that we may make in connection herewith) contains forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995 with respect to Kiniksa Pharmaceuticals, Ltd. (and its consolidated subsidiaries, collectively, unless context otherwise requires, "Kiniksa," "we," "us" or "our"). In some cases, you can identify forward looking statements by terms such as "may," "will," "should," "expect," "plan," "anticipate," "could," "intend," "goal," "design," "target," "project," "contemplate," "believe," "estimate," "predict," "potential," "strategy," or "continue" or the negative of these terms or other similar expressions, although not all forward-looking statements contain these identifying words. All statements contained in this presentation that do not relate to matters of historical fact should be considered forward-looking statements, including without limitation, statements regarding our strategy; potential value drivers; potential indications; potential market opportunities and competitive position; ongoing, planned and potential clinical trials and other studies; timing and potential impact of clinical data; regulatory and other submissions, applications and approvals; commercial strategy and commercial activities; expected run rate for our cash, cash equivalents and short-term investments; expected funding of our operating plan; financial guidance; third-party collaborations and licensing; and capital allocation.

These statements involve known and unknown risks, uncertainties, and other important factors that may cause our actual results, performance or achievements to be materially different from those expressed or implied by the forward-looking statements, including, without limitation, potential delays or difficulties with our clinical trials; potential inability to demonstrate safety or efficacy or otherwise producing negative, inconclusive or uncompetitive results; potential for changes in final data from preliminary or interim data; potential inability to replicate in later clinical trials positive results from earlier trials and studies; our reliance on third parties for manufacturing and conducting clinical trials, research and other studies; risks arising from our technology transfer of ARCALYST drug substance manufacturing; our ability to realize value from our licensing and collaboration arrangements; our ability to source sufficient drug product, as needed, to meet our clinical and commercial requirements; our inability to demonstrate safety and efficacy to the satisfaction of applicable regulatory authorities; potential for applicable regulatory authorities to not accept our filings or to delay or deny approval of any of our product candidates or to require additional data or trials to support any such approval or authorization; delays, difficulty or inability to successfully execute on our commercial strategy for ARCALYST; potential changes in our strategy, clinical trial priority, operating plan, business development strategy or funding requirements; raw materials, important ancillary product and drug substance and/or drug product shortages; substantial new or existing competition; risks arising from political and economic instability; and our ability to attract and retain qualified personnel.

These and the important factors discussed in our filings with the U.S. Securities and Exchange Commission, including under the caption "Risk Factors" contained therein could cause actual results to differ materially from those indicated by the forward-looking statements made in this presentation. These forward-looking statements reflect various assumptions of Kiniksa's management that may or may not prove to be correct. No forward-looking statement is a guarantee of future results, performance, or achievements, and one should avoid placing undue reliance on such statements. Except as otherwise indicated, this presentation speaks as of the date of this presentation. We undertake no obligation to update any forward-looking statements, whether as a result of new information, future events or otherwise, except as required by law.

This presentation also may contain estimates, projections, and/or other information regarding our industry, our business and the markets for certain of our product candidates, including data regarding the estimated size of those markets, and the incidence and prevalence of certain medical conditions. Unless otherwise expressly stated, we obtained such industry, business, market and other data from reports, research surveys, clinical trials, studies and similar data prepared by market research firms and other third parties, from industry, medical and general publications, and from government data and similar sources. Information that is based on estimates, forecasts, projections, market research, or similar methodologies is inherently subject to uncertainties and actual events or circumstances may differ materially from events and circumstances reflected in this information.

KINIKSA

Portfolio of Immune-Modulating Assets

Program	Target	Preclinical	Phase 1	Phase 2	Phase 3	Commercial
CARDIOVASCULAR FRANCHISE						
ARCALYST® (rilonacept)[1,2,3] IL-1α & IL-1β	*Recurrent Pericarditis*					
Mavrilimumab[4] GM-CSFRα	*Evaluating Potential Partnership Opportunities*					
AUTOIMMUNE FRANCHISE						
Abiprubart (KPL-404) CD40/CD154	*Rheumatoid Arthritis*					

Program	Licensee	Exclusive Licensed Territory
OUT-LICENSING AGREEMENTS		
ARCALYST (rilonacept) IL-1α & IL-1β	*Huadong Medicine*	*Asia Pacific Region, Excluding Japan*
Mavrilimumab GM-CSFRα	*Huadong Medicine*	*Asia Pacific Region, Excluding Japan*
Vixarelimab OSMRβ	*Roche and Genentech*	*Worldwide*

1) Approved in the U.S.; ARCALYST is also approved in the U.S. for cryopyrin-associated periodic syndromes (CAPS) and deficiency of the interleukin-1 receptor antagonist (DIRA); 2) The FDA granted Breakthrough Therapy designation to ARCALYST for recurrent pericarditis in 2019; the FDA granted Orphan Drug exclusivity to ARCALYST in March 2021 for the treatment of recurrent pericarditis and reduction in risk of recurrence in adults and pediatric patients 12 years and older. The European Commission granted Orphan Drug designation to ARCALYST for the treatment of idiopathic pericarditis in 2021; 3) Kiniksa has worldwide rights, excluding the Middle East and North Africa; Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan; 4) Phase 2 clinical trials of mavrilimumab in rheumatoid arthritis and giant cell arteritis achieved their primary and secondary endpoints with statistical significance; Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan
IL-1α = interleukin-1α; IL-1β = interleukin-1β; GM-CSFRα = granulocyte macrophage colony stimulating factor receptor alpha; OSMRβ = oncostatin M receptor beta

KINIKSA

ARCALYST ®

Arcalyst (rilonacept) For Injection

IL-1α AND IL-1β CYTOKINE TRAP

DISEASE AREA: Recurrent pericarditis[1]; painful and debilitating auto-inflammatory cardiovascular disease

COMPETITION[2]: First and only FDA-approved therapy for recurrent pericarditis

REGULATORY: U.S. Orphan Drug exclusivity for treatment of and reduction in risk of recurrence of recurrent pericarditis; European Commission Orphan Drug designation in idiopathic pericarditis

STATUS: FDA-Approved

ECONOMICS: 50/50 split on profit and third-party proceeds

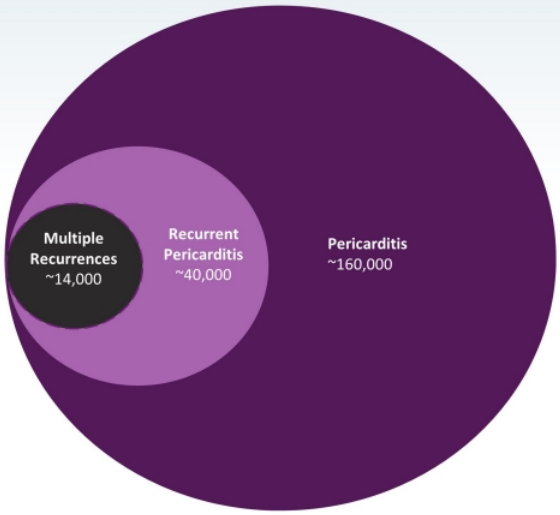
RIGHTS: Kiniksa has worldwide rights[3] (excluding MENA) for all indications outside those in oncology and local administration to the eye or ear

KINIKSA

1) ARCALYST is also approved and marketed for Cryopyrin-Associated Periodic Syndromes (CAPS) and maintenance of remission of Deficiency of Interleukin-1 Receptor Antagonist (DIRA) in the United States;
2) Drugs@FDA: ARCALYST Prescribing Information, Ilaris Prescribing Information, Kineret Prescribing Information; Kaiser et al. Rheumatol Int (2012) 32:295–299; Theodoropoulou et al. Pediatric Rheumatology 2015, 13(Suppl 1):P155; Fleischmann et al, 2017 ACR/ARHP Abstract 1196; Kosloski et al, J of Clin Pharm 2016, 56 (12) 1582-1590; Cohen et al. Arthritis Research & Therapy 2011, 13:R125; Cardiel et al. Arthritis Research & Therapy 2010, 12:R192; Hong et al. Lancet Oncol 2014, 15: 656-666; 3) Kiniksa granted Huadong Medicine exclusive rights in the Asia Pacific Region, excluding Japan;
IL-1α = interleukin-1α ; IL-1β = interleukin-1β; MENA = Middle East North Africa

Pericarditis Epidemiology

Of the 14,000 target population with multiple recurrences there is a high turnover of ~50% of patients each year, meaning ongoing opportunities to ensure diagnosis and targeted treatment



All figures annual period prevalence

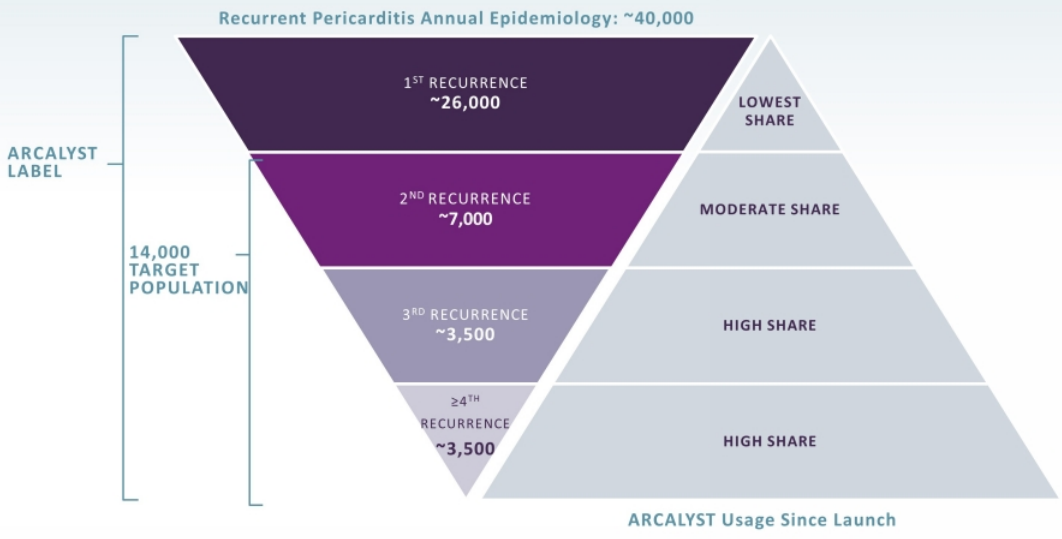
Approximately 14,000 recurrent pericarditis patients in the U.S. suffer from persistent underlying disease, with multiple recurrences and inadequate response to conventional therapy[1]

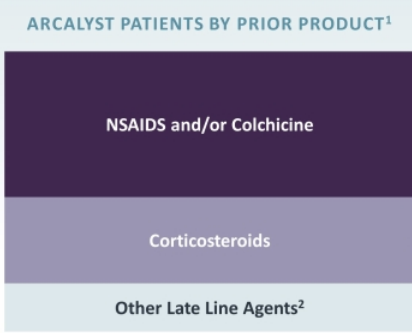
- **~160,000:** Epidemiological analysis using large national surveillance databases to calculate the pooled annualized prevalence of pericarditis ***(Basis for Orphan Drug Designation approval)[2]***
- **~40,000:** Up to 30% experience at least one recurrence; some recur over multiple years[6,7]
- **~14,000:** Nearly 50% annual turnover with ~7,000 patients entering into the pool each year[8]

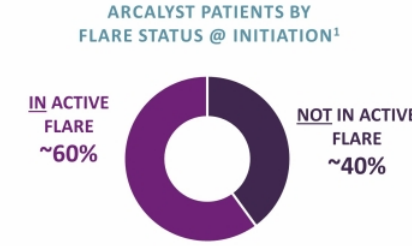
KINIKSA

1) Cremer et al. American Journal of Cardiology. 2016;2311-2328; 2) DOF, Kiniksa Pharmaceuticals, Ltd.; 3) Brucato A, Maestroni S, Cumetti D, et al. Autoimmun Rev. 2008; 8:44-47; 4) Lange R, Hills L. N Engl J Med. 2004; 351: 2195-2202; 5) Imazio M, Cecchi E, Demichelis B, et al. Circulation. 2007; 115: 2739-2744; 6) Imazio et al. Circulation. 2005;112:2012-2016; 7) Adler et al. Circulation. 1998;97:2183-2185; 8) Klein A, Cremer P, Kontzias A, et al. US database study of clinical burden and unmet need in recurrent pericarditis. J Am Heart Assoc. 2021; 10:e018950. doi:10.1161/JAHA. 120.018950

Treated Patients Since Launch Are Closely Associated to the 14,000 Target Population, While Prescribers Can Utilize ARCALYST Earlier in the Disease





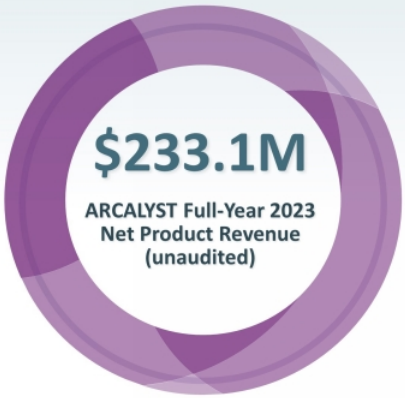


Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Magestro M. Annals of Epidemiology. 2019;36:71; 2) Lin D, Majeski C, DerSarkissian M, Magestro M, Cavanaugh C, Laliberte F, Lejune D, Mahendran M, Duh M, Klein A, Cremer P, Kontzias A, Furqan M, Tubman R, Roy M, Mage. (Nov, 2019). Real-World Clinical Characteristics and Recurrence Burden of Patients Diagnosed with Recurrent Pericarditis in the United States. Poster session presented at the American Heart Association, Philadelphia, PA.; 3) ClearView Forecasting Analysis 2019 Q1

Source: 1) Kiniksa Pharmaceuticals data on file 2023. 2) Other late line agents include anakinra, azathioprine, methotrexate

ARCALYST Commercial Growth in 2023: By the Numbers

~78% annual growth vs Q4 2022



~90% annual growth vs full-year 2022

~80% annual growth vs end of Q4 2022

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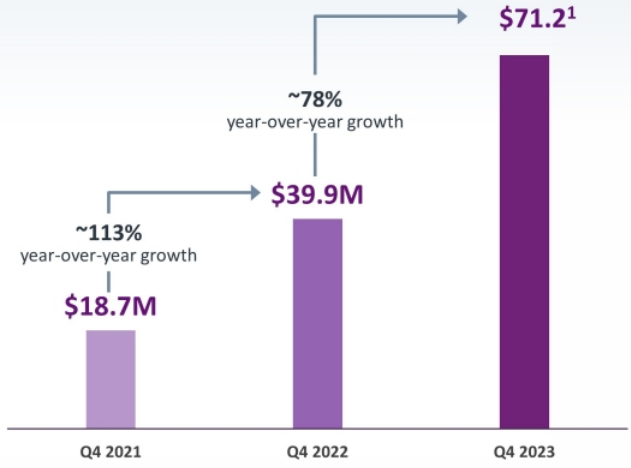
Strong Q4 2023 ARCALYST Net Product Revenue Growth

Total Net Revenue Growth per Quarter



Total Prescribers	**>1,700**
Repeat Prescribers (% of Total)	**~24%**
Payer Approval (% of Completed Cases)	**>90%**
Average Total Duration of Therapy	**~23 months**
Patient Compliance	**>85%**

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1) ARCALYST net product revenue (unaudited)

Key Executional Priorities to Drive Greater Patient and Physician Adoptio

Identify appropriate patients and drive a proactive mindset with physicians and patients

Close the ARCALYST **knowledge gap** with physicians

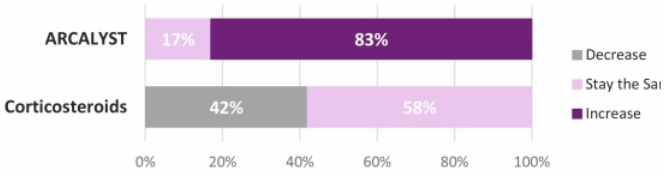
Advance the **treatment paradigm**

Educate on **duration of disease** and treatment

Externally: Thought leaders are introducing treatment paradigms for recurrent pericarditis that recommend IL-1 antagonists, such as ARCALYST, be used ahead of corticosteroids[1]

Our Aim: Continue to drive the evolution of this treatment paradigm

Intended Future Use Among Target Healthcare Providers[2]



- Of target physicians who have knowledge of ARCALYST, they overwhelmingly expect to **increase their prescribing of ARCALYST in next 6 months**
- The biggest barriers for physicians to prescribing ARCALYST are **limited knowledge about the product and/or experience with the payer approval process**

1) Dong, Klein, Wang. Paradigm Shift in Diagnosis and Targeted Therapy in Recurrent Pericarditis. Springer Nature. 2023.; Klein, Cremer, Kafil. Recurrent Pericarditis A Promising Future for IL-1 Blockers in Autoinflammatory Phenotypes. Journal of the American College of Cardiology, Editorial Comment. 2023.; Thomas, Bonaventura, Vecchié, et al. Interleukin-1 blockers for the treatment of recurrent pericarditis: pathophysiology, patient reported outcomes and perspectives. Journal of Cardiovascular Pharmacology. 2023.; Imazio, Mardigyan, Andreis, et al. New developments in the management of recurrent pericarditis. Canadian Journal of Cardiology. 2023.; Kumar, Khubber, Reyaldeen, et al. Advances in Imaging and Targeted Therapies for Recurrent Pericarditis. JAMA Cardiology Review. 2022.; Sushil, Cremer, Raisinghani.
2) HCP Market Research, Q3 2023; Kiniksa Data on File.

Average Total Duration of ARCALYST Therapy: ~23 Months[1]

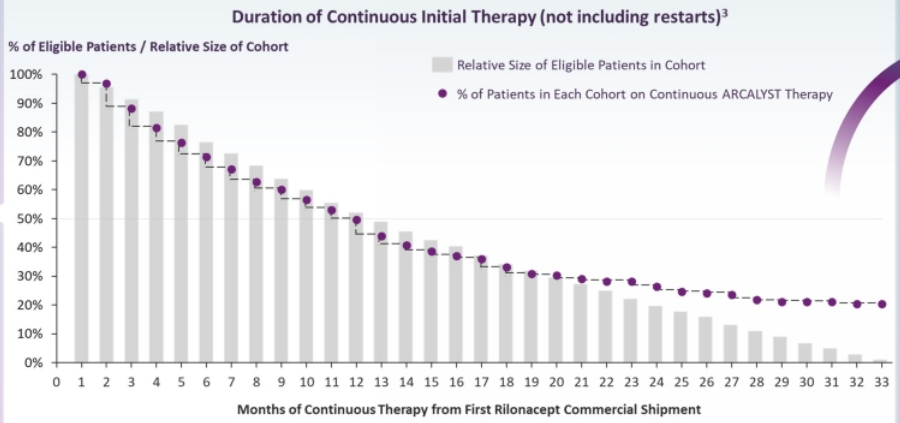
Advancing the treatment paradigm to treat continuously throughout disease duration (median 3 years[2])

Average ***Initial*** Duration of Therapy

~15 Months[1]

Median ***Initial*** Duration of Therapy

~12 Months[1]



~45%

Of Patients Restarted Therapy Following Initial Discontinuation

(Within ~8 weeks)

~23 Months Average ***Total*** Duration of Therapy After Accounting for Patient Restarts

1) As of Q4 2023; 2) Lin D, Laliberté F, Majeski C, et al. Disease and economic burden associated with recurrent pericarditis in a privately insured United States population. Adv Ther. 2021;38(10):5127-5143. doi:10.1007/s12325-021-01868-7; 3) Initial continuous therapy is determined to have ended if greater than 28 days elapses beyond the exhaustion date of a patient's most recent days supplied without an observed refill of ARCALYST

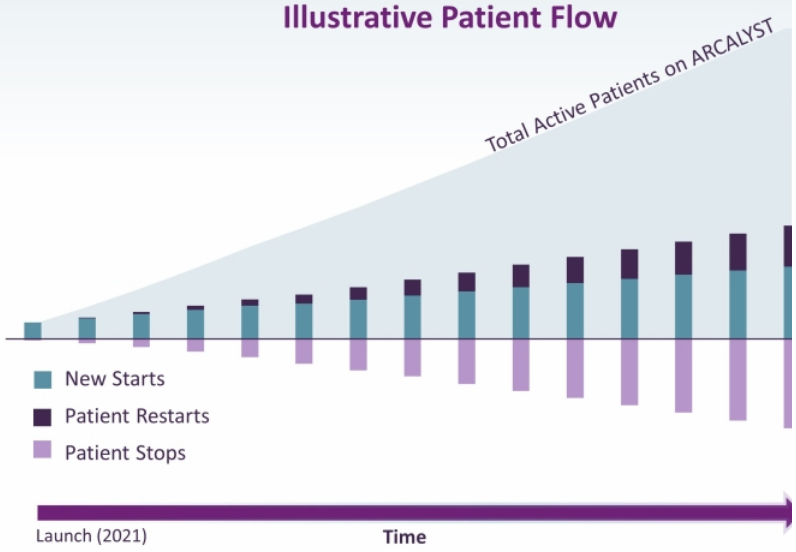
Growth in Total Patients on ARCALYST Therapy

Acceleration in new-to-brand and restart patients offset higher patient stops over time

ARCALYST Patient Flow

New to Brand Patients	Strong sequential growth: >1,700 unique prescribers; ~24% of which are repeat prescribers
−	
Patient Stops	Increases over time as base of active ARCALYST patients grows with Initial Starts and Restarts
+	
Patient Restarts	Increases over time as patient stops increase; currently ~45% after ~8 weeks
=	
Active Patients	Increases over time driven by new-to-brand and restart growth; as of Q4 2023, ~9% of 14K multiple recurrence target patient population

Illustrative Patient Flow



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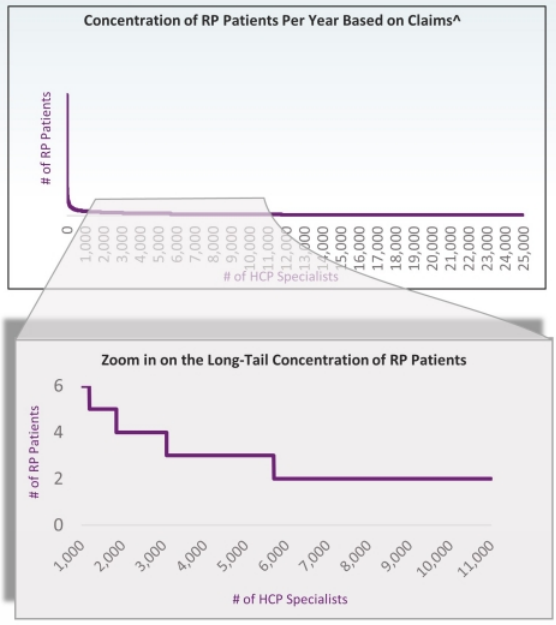
Evolving ARCALYST Field Strategy

Targeting an increased number of top and mid-tier physicians

The recurrent pericarditis population is widely dispersed



Data driven expansion to field sales team

Q4 2022	Q4 2023
Prior expansion to create greater reach & frequency	New expansion to provide greater frequency on top tier physicians and improved coverage to the mid tier
~50 Specialty Cardiology Reps	**~85** Specialty Cardiology Reps
Reaching:	Reaching:
~6,000 top and mid tier prescribers	**~11,000** top and mid tier prescribers
~70%* of RP patients nationally	**~85%*** of RP patients nationally

- In any given year, the 14,000 multiple recurrent pericarditis patients may present to any of the >20,000 cardiologists and >5,000 rheumatologists in US
- With our field expansion, we expect to accelerate coverage and frequency among the top tier as well as the long tail of physicians who may identify recurrent pericarditis patients
  - Data-driven decisions ensured continued growth in collaboration profitability following the prior expansion
  - With the new expansion, we have the opportunity to meaningfully increase frequency on prior field targets and to reach new health care providers that have no prior field interactions

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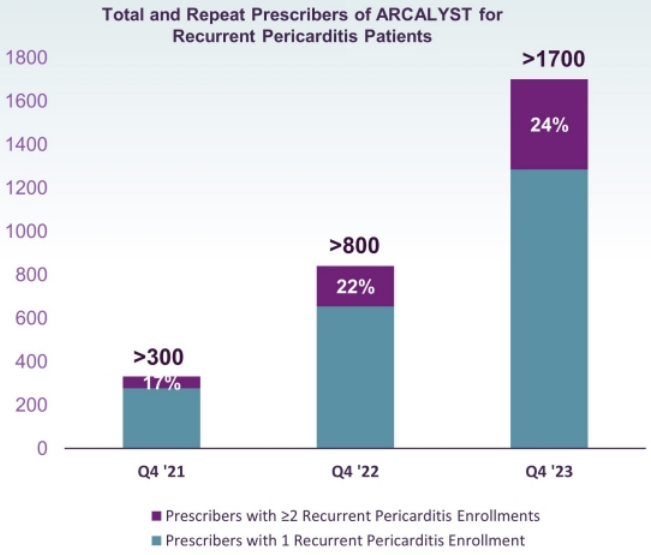
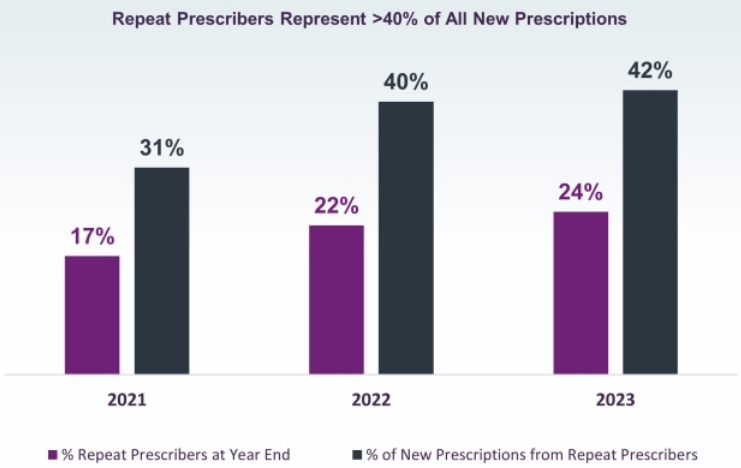
*Including targets, prospects, and opportunistic calls to non-targets

^Internal analysis based on Komodo Claims Data; includes patients with at least 1 recurrence

Opportunity for Continued ARCALYST Growth Remains High





The increase in repeat prescribers has continued to grow against the backdrop of a rapidly increasing total prescriber base (>300 at YE 2021 vs >1,700 at YE 2023)

As of the end of 2023, ~9% of the target 14,000 recurrent pericarditis patients with multiple-recurrences were actively on ARCALYST treatment

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Pricing, Access and Distribution Considerations

Pricing

- ARCALYST list price of $22,603 per month

  Based on first and only FDA-approved therapy for recurrent pericarditis, in-line with specialty biologics with Breakthrough Therapy and Orphan Drug designation

- Helping to ensure **patient affordability** and access to treatment is one of our core principles and to this end, we offer a suite of programs to support affordability to eligible patients who are prescribed ARCALYST; eligible patients are able to get ARCALYST for a copay of as low as $0

Access

- Kiniksa's goal is to maintain rapid and broad access to ARCALYST for patients with Recurrent Pericarditis, CAPS, and DIRA
- Payer mix for ARCALYST is largely **commercial (~70%), Medicare (~20%), Medicaid (~10%)**
- Payer engagement has increased awareness of recurrent pericarditis and the differentiated value of ARCALYST
- The **Kiniksa OneConnect™** program is a personalized treatment support program for patients prescribed ARCALYST

Distribution

- ARCALYST is distributed **through a closed network of designated specialty pharmacies and the Veterans Affairs**
- The distribution network for ARCALYST was developed to provide a high and consistent level of patient support with broad access. Network pharmacies provide customized services to support patients

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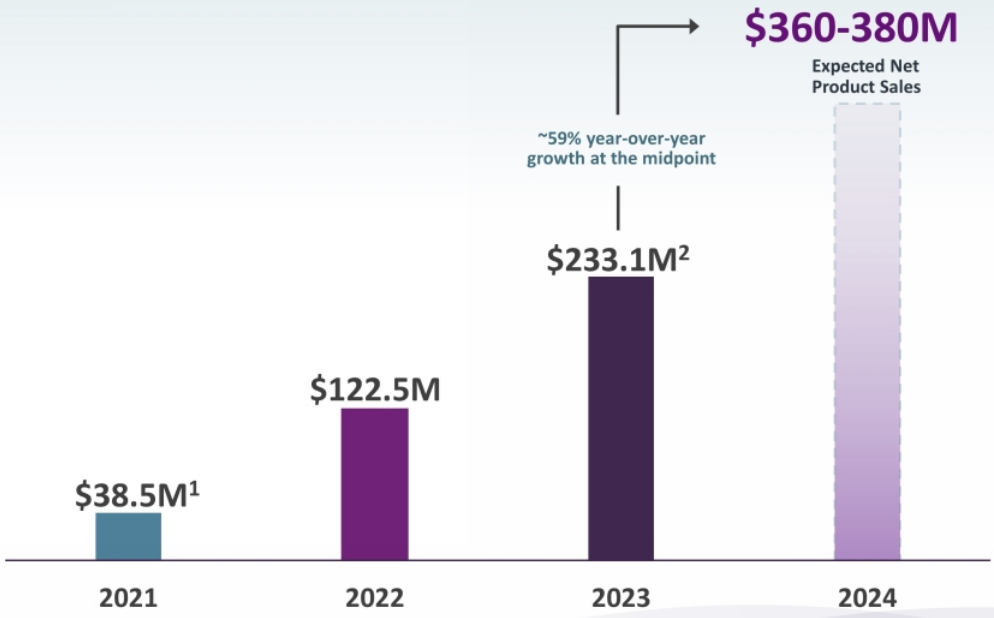
CAPS = Cryopyrin-Associated Periodic Syndromes ; DIRA = Deficiency of IL-1 Receptor Antagonist

2024 ARCALYST Net Product Revenue Guidance

Well-positioned to expand the breadth and depth of ARCALYST in recurrent pericarditis



1) 9 Months of Commercial Availability; 2) ARCALYST net product revenue (unaudited)

Summary of ARCALYST Profit Share Arrangement with Regeneron[1]

- Kiniksa is responsible for sales and distribution of ARCALYST in all approved indications in the United States.
- Kiniksa's license to ARCALYST includes worldwide rights*, excluding MENA, for all applications other than those in oncology and local administration to the eye or ear.
- Kiniksa covers 100% of development expenses related to approval of additional indications.
- Kiniksa evenly splits profits on ARCALYST sales and licensing proceeds with Regeneron

1) Subject to description contained in definitive agreement; 2) Global net sales for CAPS, DIRA and recurrent pericarditis recognized as revenue on Kiniksa's income statement; 3) Profit Split-Eligible Cost of Goods Sold = total cost of goods sold - amortization of Regeneron milestone payment
*Kiniksa exclusively licensed rights for the development and commercialization of ARCALYST in APAC (ex-Japan) to Huadong Medicine
CAPS = Cryopyrin-Associated Periodic Syndromes; DIRA = Deficiency of the Interleukin-1 Receptor Antagonist; MENA =Middle East and North Africa; APAC = Asia Pacific Region

ABIPRUBART (KPL-404)

ANTI-CD40 MONOCLONAL ANTIBODY INHIBITOR OF THE CD40-CD154 COSTIMULATORY INTERACTION

DISEASE AREA: Rheumatoid Arthritis; a chronic inflammatory disorder affecting many joints; External proof-of-concept previously established in broad range of autoimmune diseases: Sjogren's disease, systemic lupus, solid organ transplant and Graves' disease[1,2]

SCIENTIFIC RATIONALE[3,4]: Attractive target for blocking T-cell dependent, B-cell–mediated autoimmunity

STATUS: Phase 2 proof-of-concept study of chronic subcutaneous administration ongoing; data from Cohort 4 expected in Q2 2024

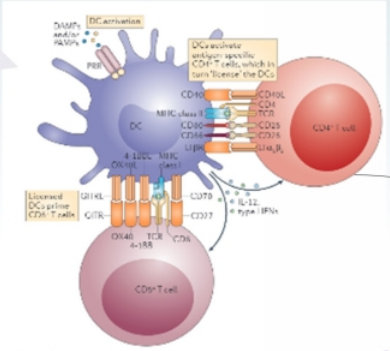
ECONOMICS: Negligible clinical and regulatory milestones and royalty on annual net sales

RIGHTS: Worldwide

Sources: 1) Muralidharan et al. Preclinical immunopharmacologic assessment of KPL-404, a novel, humanized, non-depleting antagonistic anti-CD40 monoclonal antibody. J Pharmacol Exp Ther. 2022, 381(1):12-21. 2) Samant M, Ziemniak J, Paolini JF. First-in-Human Phase 1 Randomized Trial with the Anti-CD40 Monoclonal Antibody KPL-404: Safety, Tolerability, Receptor Occupancy, and Suppression of T-Cell-Dependent Antibody Response. J Pharmacol Exp Ther. 2023 Dec;387(3):306-314. 3) Elgueta, et al. Immunol Rev 2009, 229 (1), 152-172; 4) Peters, et al. Semin Immunol 2009, 21 (5) 293-300
RO = receptor occupancy; TDAR = T-cell Dependent Antibody Response

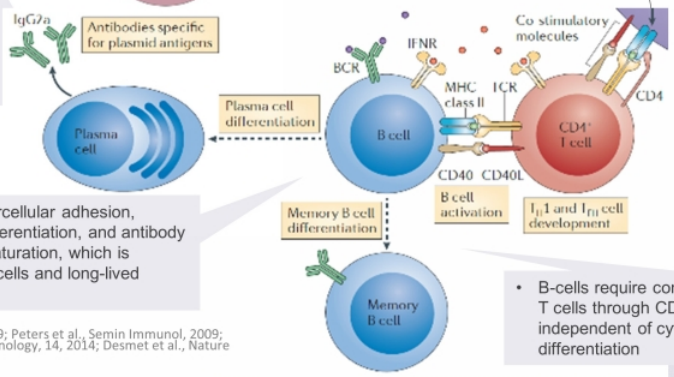
CD40/CD154 Interaction: Essential Immune Pathway for T-Cell Priming and T-Cell Dependent B-Cell Responses

- CD40 is expressed on the surface of dendritic cells, B-cells, antigen-presenting cells and non-immune cell types
- Its ligand, CD40L (CD154), is expressed by activated T-cells, platelets, and other cell types



- CD40 ligation on DCs induces cell maturation by promoting antigen presentation and enhancing their costimulatory activity
- Mature DCs stimulate activated T-cells to increase IL-2 production that facilitates T-helper cells (Th) and cytolytic T-Lymphocyte (CTL) expansion
- CD40-stimulated DCs also secrete cytokines favoring Th1 cell differentiation and promoting Th cell migration to sites of inflammation
- CD40 ligation also provides a pro-inflammatory signal within the mononuclear phagocyte system

- Humoral immunity is dependent on a thriving B cell population and activation by Th cells; blockade of CD40/CD40L interaction has been shown to completely ablate primary and secondary TDAR response



- CD40 engagement triggers B-cell intercellular adhesion, sustained proliferation, expansion, differentiation, and antibody isotype switching leading to affinity maturation, which is essential for generation of memory B cells and long-lived plasma cells

- B-cells require contact-dependent stimulus from T cells through CD40/CD40L interaction independent of cytokines to trigger growth and differentiation

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Sources: Elgueta et al., Immunol Rev, 2009; Peters et al., Semin Immunol, 2009; Kambayashi et al., Nature Reviews: Immunology, 14, 2014; Desmet et al., Nature Reviews: Immunology, 12, 2012

Abiprubart Phase 2 Trial in Rheumatoid Arthritis

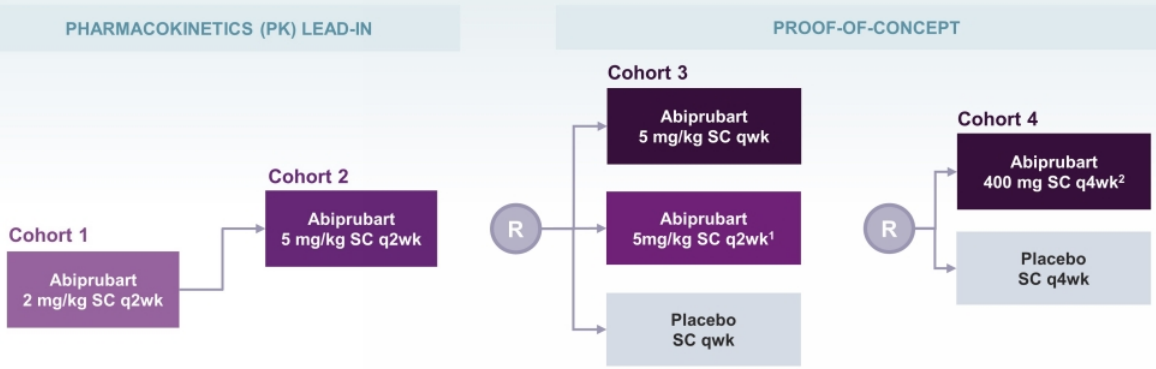
Study to evaluate the efficacy, dose response, PK, and safety of chronic SC dosing over a 12-week treatment durati

PATIENT POPULATION:

- Patients with active RA who have been treated with a biological disease-modifying anti-rheumatic drug (bDMARDs) AND/OR Janus kinase inhibitor (JAKi) therapy for RA for ≥ 3 months and who have had inadequate response or have had to discontinue bDMARD and/or JAKi therapy due to intolerance or toxicity, regardless of treatment duration.

DISEASE CRITERIA:

- Six or more swollen joints and ≥ 6 tender joints at screening and baseline line visits; levels of high sensitivity C-reactive protein ≥ 5 mg/L; seropositivity for serum RF and/or ACPA at screening.



PK Lead-In: Cohorts 1-2

- Each cohort sequentially randomized 8 patients in a 3:1 (active:placebo) ratio; placebo recipients from Cohorts 1 and 2 were pooled
- Primary Endpoints:
  - Incidence of treatment-emergent adverse events (TEAEs)
  - Pharmacokinetics (C_{max}, $AUC_{(0\text{-}t)}$)
- Secondary Efficacy Endpoint:
  - Change from baseline in DAS28-CRP at Week 12

Proof of Concept: Cohorts 3-4

- Cohort 3 randomized 78 patients in a 1:1:1 ratio (n~26/arm)
- Cohort 4 randomized 51 patients in a 3:2 ratio (n=~20-30/arm)
- Primary Efficacy Endpoint:
  - Change from baseline in DAS28-CRP at Week 12
- Secondary Endpoints :
  - Incidence of treatment-emergent adverse events (TEAEs)
  - Pharmacokinetics (C_{max}, $AUC_{(0\text{-}t)}$)

1) The 5 mg/kg SC q2wk group will receive weekly administrations of alternating active investigational product and matching blinded placebo
2) The Cohort 4 Abiprubart 400mg SC q4wk group includes a 600mg loading dose on Day 1

SC = subcutaneous; qwk = every week; q2wk = every other week; q4wk = every four weeks; AUC = Area Under the Curve; RF = Rheumatoid Factor; ACPA = anti-citrullinated protein antibodies, PD = Pharmacodynamics; PK = Pharmacokinetics; R = Randomization

Baseline Demographics (Cohort 3)[1]

	Abiprubart 5mg/kg SC qwk (n=27)	Abiprubart 5mg/kg SC q2wk (n=25)	Placebo (n=26)	Total (n=78)
Mean Age (Years)	58.5	60.0	57.6	58.7
Sex % (Male/Female)	18.5/81.5	20.0/80.0	7.7/92.3	15.4/84.6
Race				
White %; (n)	92.6 (n=25)	92.0 (n=23)	92.3 (n=24)	92.3 (n=72)
Black or African American %; (n)	3.7 (n=1)	8.0 (n=2)	7.7 (n=2)	6.4 (n=5)
Asian %; (n)	3.7 (n=1)	0	0	1.3 (n=1)
Country[2]				
United States %; (n)	29.6 (n=8)	28.0 (n=7)	38.5 (n=10)	32.1 (n=25)
Bulgaria %; (n)	0	4.0 (n=1)	11.5 (n=3)	5.1 (n=4)
Czechia %; (n)	11.1 (n=3)	4.0 (n=1)	3.8 (n=1)	6.4 (n=5)
Georgia %; (n)	7.4 (n=2)	12.0 (n=3)	11.5 (n=3)	10.3 (n=8)
Hungary %; (n)	18.5 (n=5)	4.0 (n=1)	3.8 (n=1)	9.0 (n=7)
Poland %; (n)	25.9 (n=7)	28.0 (n=7)	19.2 (n=5)	24.4 (n=19)
South Africa %; (n)	7.4 (n=2)	20.0 (n=5)	11.5 (n=3)	12.8 (n=10)

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1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing; 2) Cohorts 1 and 2 were conducted entirely in the United States

Baseline Disease Characteristics were Balanced Across Treatment Arms (Cohort 3)[1]

	Abiprubart 5mg/kg SC qwk (n=27)	Abiprubart 5mg/kg SC q2wk (n=25)	Placebo (n=26)	Total (n=78)
DAS28-CRP Score				
DAS28-CRP[2]	5.58	5.92	5.98	5.82
Tender Joint Count-28[2]	13.4	16.1	15.4	14.9
Swollen joints-28 (mean)	10.1	12.2	12.0	11.4
Patient Global Assessment[2]	6.68	6.49	6.73	6.64
C-Reactive Protein (mg/L)[2]	16.00	18.72	26.74	20.45
Mean Duration of Rheumatoid Arthritis (years)	12.24	13.50	15.47	13.72
Rheumatoid factor (IU/mL)[2]	165.21	183.45	154.62	167.53
Anti-Cyclic Citrullinated Peptide %; (n)				
Positive	74.1 (n=20)	80.0 (n=20)	76.9 (n=20)	76.9 (n=60)
Negative	22.2 (n=6)	20.0 (n=5)	23.1 (n=6)	21.8 (n=17)
Intermediate	3.7 (n=1)	0	0	1.3 (n=1)

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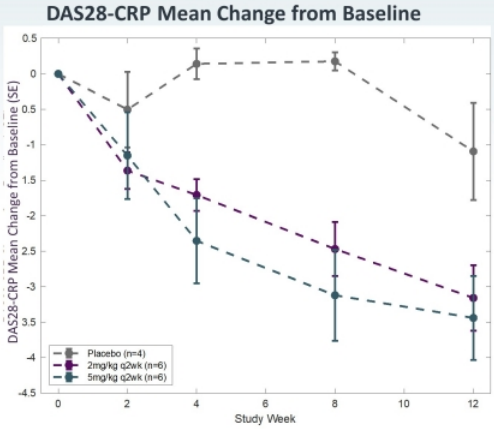
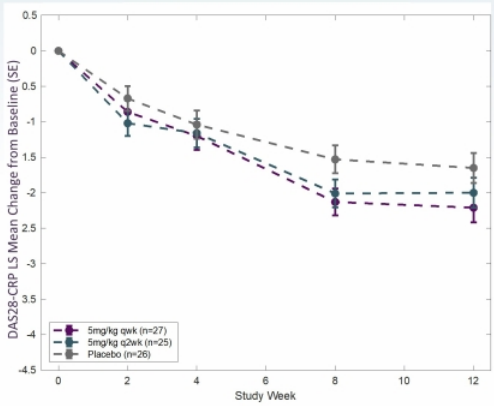
1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing; 2) Mean

Phase 2 Clinical Trial of Abiprubart in Rheumatoid Arthritis Met Primary Efficacy Endpoint (Change from Baseline in DAS28-CRP vs Placebo at Week 12)

Cohorts 1 & 2
DAS28-CRP Mean Change from Baseline



Cohort 3
DAS28-CRP LS Mean Change from Baseline[1]



Cohort 1: in the abiprubart 2 mg/kg SC biweekly dosing group (n=6), mean change from baseline in DAS28-CRP at Week 12 was -3.16 points compared to -1.09 points in pooled placebo recipients (n=4), (Mean Difference = -2.07, p=0.0312)

Cohort 2: in the abiprubart 5 mg/kg SC biweekly dosing group (n=6), mean change from baseline in DAS28-CRP at Week 12 was -3.44 points compared to -1.09 points in pooled placebo recipients (n=4), (Mean Difference = -2.35, p=0.0338)

In the abiprubart 5 mg/kg SC weekly dosing group (n=27), LS mean change [95% (CI)] from baseline in DAS28-CRP at Week 12 was -2.21 [-2.62, -1.80] points compared to -1.65 [-2.07, -1.23] points in placebo recipients (n=26), (LS Mean Difference = -0.56, p=0.0487)

In the abiprubart 5 mg/kg SC biweekly dosing group (n=25), LS mean change [95% CI] from baseline in DAS28-CRP at Week 12 was -2.00 [-2.43, -1.58] points compared to -1.65 [-2.07, -1.23] points in placebo recipients (n=26), (LS Mean Difference = -0.35, p=0.2140)

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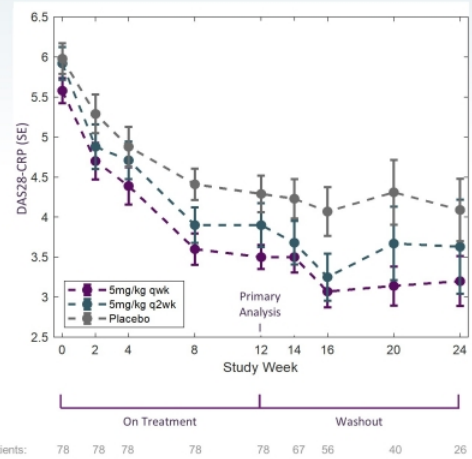
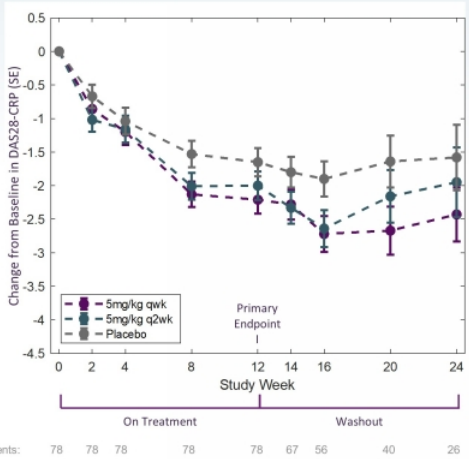
1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing;
DAS28-CRP = Disease Activity Score of 28 Joints Using C-reactive Protein; SC = Subcutaneous; LS = Least Squares; CI = Confidence Interval

DAS28-CRP Scores Over Time (Cohort 3) [1]

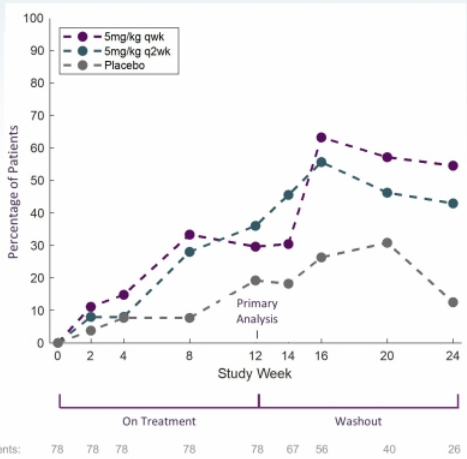
DAS28-CRP Score Through Week 24



DAS28-CRP LS Mean Change from Baseline Through Week 24

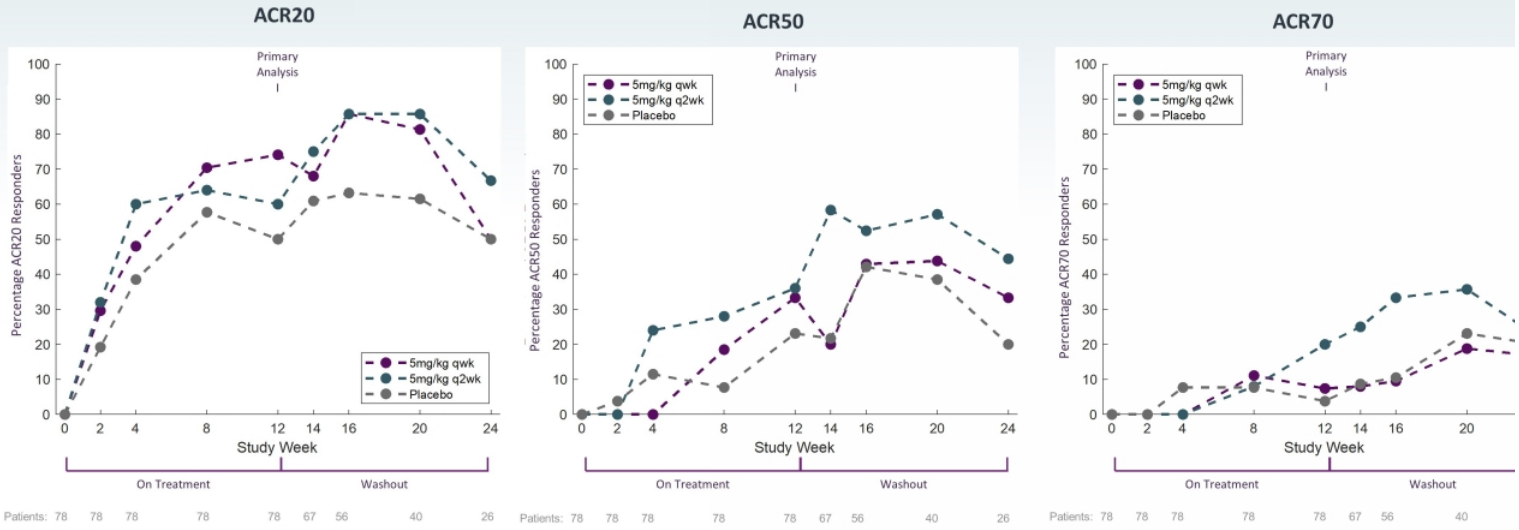


Patients Achieving DAS28 Low Disease Activity or Remission



1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing;
DAS28-CRP = Disease Activity Score of 28 Joints Using C-reactive Protein; Low Disease Activity = patients achieving DAS28-CRP low disease activity (≥2.6 and < 3.2); Remission = patients achieving DAS28-CRP remission (< 2.6)

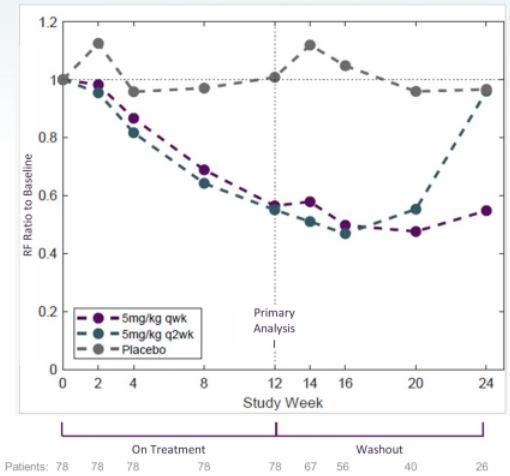
ACR Responders Over Time (Cohort 3)[1]



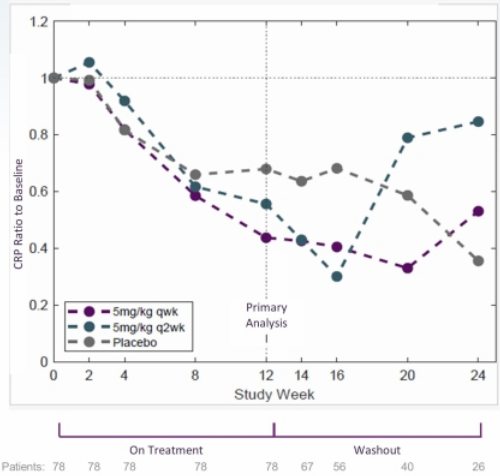
1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing;
ACR20 = a composite measure defined as an improvement of 20% in the number of tender and swollen joints and a 20% improvement in three of the following five criteria: patient global assessment, physician global assessment, functional ability measure [most often Health Assessment Questionnaire (HAQ)], visual analog pain scale, and erythrocyte sedimentation rate or C-reactive protein (CRP); ACR50 and ACR70 = the same instruments as ACR20 with improvement levels defined as 50% and 70%, respectively, versus 20% for ACR20.

Abiprubart Significantly Reduced Disease-Related Inflammatory Markers (Cohort 3)[1]

Rheumatoid Factor
Geometric Mean Ratio to Baseline



C-Reactive Protein
Geometric Mean Ratio to Baseline



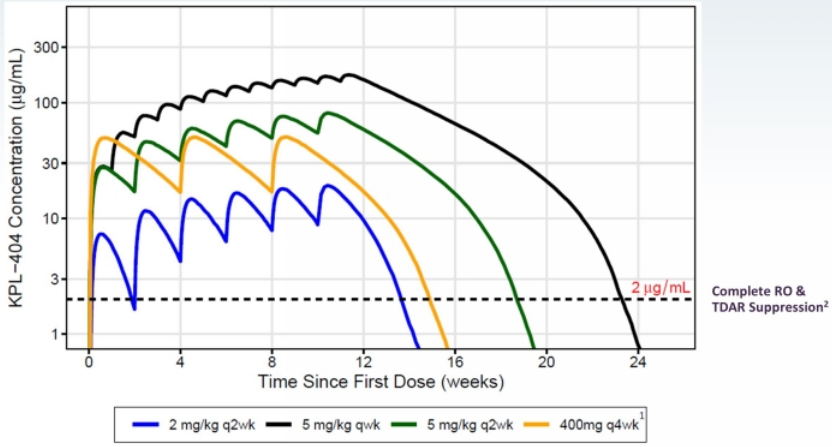
1) Modified Intention to Treat (mITT) analysis population (all randomized subjects who received at least one dose of study drug and had a baseline assessment and at least one post-baseline assessment for the primary efficacy endpoint); the Phase 2 study of abiprubart in rheumatoid arthritis is ongoing, this topline analysis includes all patients having reached Week 12, and follow-up to Week 24 is ongoing

Abiprubart was Well-Tolerated in Phase 2 RA Trial (Cohort 3 Data)[1]

Category[2]	Abiprubart 5mg/kg SC qwk (n=27)	Abiprubart 5mg/kg SC q2wk (n=25)	Placebo (n=26)
Treatment Emergent Adverse Events (TEAEs)[3]	48.1 (n=13)	28.0 (n=7)	30.8 (n=8)
Drug Related TEAE[4]	7.4 (n=2)	8.0 (n=2)	7.7 (n=2)
TEAEs by Maximum severity[5]	48.1 (n=13)	28.0 (n=7)	30.8 (n=8)
Mild	33.3 (n=9)	16.0 (n=4)	15.4 (n=4)
Moderate	14.8 (n=4)	12.0 (n=3)	15.4 (n=4)
Severe	0	0	0
Potentially Life Threatening	0	0	0
Fatal	0	0	0
Serious TEAEs (SAE)	3.7 (n=1)[5]	0	0
Drug-Related SAEs[3]	0	0	0
TEAEs Leading to Death	0	0	0
TEAEs Leading to Dose Interruption	3.7 (n=1)	0	3.8 (n=1)
TEAEs Leading to Treatment Discontinuation	0	0	0
TEAEs of Special Interest	0	4.0 (n=1)	0
Injection Site Reaction	3.7 (n=1)	4.0 (n=1)	0

1) Safety Population: All randomized subjects who received at least one dose of study drug; 2) all categories are represented in percentages; 3) Defined as any event not present before exposure to study drug or any event already present that worsens in either intensity or frequency after exposure to study drug during treatment period; 4) Definitely related or possibly related, as assessed by the investigator; 5) Each subject has only been represented with the maximum severity; 5) Monaural deafness at Week 12, not related, resolved with pulse-dose steroids

PK-Modeling and Dose Simulations for Abiprubart Phase 2 RA Study



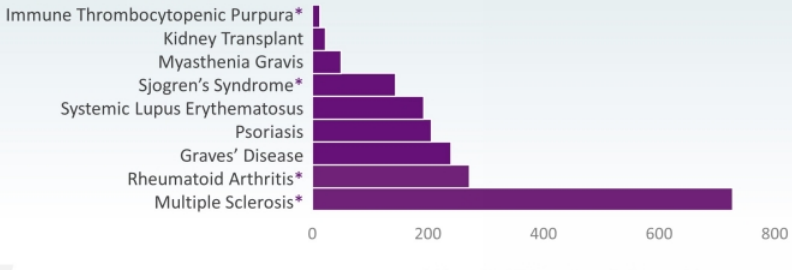
Modeling data[3] place 400 mg q4wk dose level (Cohort 4) intermediate between 2 mg/kg q2wk and 5 mg/kg q2v

1) The Cohort 4 KPL-404 400mg SC q4wk group includes a 600mg loading dose on Day 1; 2) Serum concentration predicted based on Phase 1 data; 3) PK model generated based on PK data from Cohorts 1-3 of the abiprubart Phase 2 trial in Rheumatoid Arthritis as well as Phase 1 data from healthy volunteers
RO = receptor occupancy; TDAR = T-Cell Dependent Antibody Response

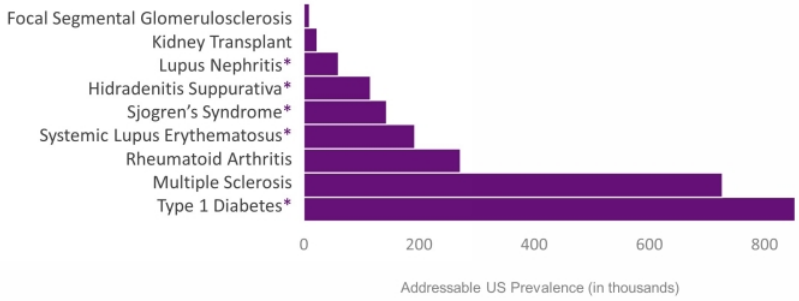
Potential for Evaluation of Abiprubart in a Range of Autoimmune Diseases

CD40/CD154 interaction has been implicated in a number of devastating diseases

Indications with Published Data



Indications with Pending Data & Trials Ongoing



INDICATION SELECTION CRITERIA

- Robust data or proof-of-concept supporting mechanism
- Differentiation vs. competitors
- Commercial attractiveness

*Indications evaluated with subcutaneous administration

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Sources: 2019 numbers: https://unos.org/data/transplant-trends/; Hunter et al. Prevalence of rheumatoid arthritis in the United States adult population in healthcare claims databases, 2004-2014; Rheumatol Int. 2017 Sep;37(9):1551-1557; Overall Prevalence: Maciel et al, Arthritis Care Res (Hoboken) 2017; Qin et al, Ann Rheum Dis 2015; UpToDate; Baldini et al. Prevalence of Severe Extra-Glandular Manifestations in a Large Cohort of Patients with Primary Sjögren's Syndrome; 2012 ACR/ARHP Annual Meeting, ABSTRACT NUMBER: 2185; Wallin et al. The prevalence of MS in the United States A population-based estimate using health claims data, Neurology, March 5, 2019; Somers et al.; Prevalence of Systemic Lupus Erythematosus in the United States: Preliminary Estimates from a Meta-Analysis of the Centers for Disease Control and Prevention Lupus Registries; 2019 ACR/ARP Annual Meeting ABSTRACT NUMBER: 2886; Garg et al. JAMA Dermatol. 2017;153(8):760-764. doi:10.1001/jamadermatol.2017.0201 Sex- and Age-Adjusted Population Analysis of Prevalence Estimates for Hidradenitis Suppurativa in the United States; MayoClinic.org; Yale J Biol Med. 2013 Jun; 86(2): 255–260. N Engl J Med 2016;375:2570-81; https://www.diabetesresearch.org/diabetes-statistics; Nephcure.org; Kitiyakara C, Eggers P, Kopp JB. Twenty-one-year trend in ESRD due to focal segmental glomerulosclerosis in the United States. Am J Kidney Dis. 2004 Nov;44(5):815-25; Rachakonda et al. J Am Acad Dermatol . 2014 Mar;70(3):512-6. doi: 10.1016/j.jaad.2013.11.013. Epub 2014 Jan 2. Psoriasis prevalence among adults in the United States; Yeung et al. Psoriasis severity and the prevalence of major medical co-morbidities: a population-based study; JAMA Dermatol. 2013 Oct 1; 149(10):1173–1179; Hoover et al. Kidney Int. 2016 Sep; 90(3): 487–492. Insights into the Epidemiology and Management of Lupus Nephritis from the U.S. Rheumatologist's Perspective.

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Financials
Third Quarter 2023

Third Quarter 2023 Financial Results

Income Statement	Three Months Ended September 30,	
	2023	2022
Product Revenue	$64.8M	$33.4M
License and Collaboration Revenue	$2.2M	$65.7M
Total Revenue	**$67.0M**	**$99.1M**
Cost of Goods Sold	$9.1M	$6.9M
Collaboration Expenses[1]	$17.3M	$4.6M
Research and Development	$17.1M	$16.5M
Selling, General and Administrative	$34.5M	$24.7M
Total Operating Expenses	**$78.0M**	**$52.7M**
Income Tax Benefit (Provision)	**$(5.4M)**	**$177.4M**
Net Income (Loss)	**$(13.9M)**	**$224.1M**

Collaboration Expenses[1]	Three Months Ended September 30	
	2023	202
ARCALYST Net Sales (RP + CAPS + DIRA)	**$64.8M**	**$33.4**
Profit Split-Eligible Cost of Goods Sold[2]	($8.8M)	($6.7M
Commercial, Marketing, Regulatory and Other Expenses	($21.4M)	($17.5M
ARCALYST Collaboration Operating Profit	**$34.6M**	**$9.2**
ARCALYST Licensing Proceeds	$0.0M	$0.0
Collaboration Expenses[1]	**$17.3M**	**$4.6**

Balance Sheet	September 30, 2023	December 3 202
Cash, Cash Equivalents and Short-term Investments	**$201.1M**	**$190.6**

Cash reserves of $206.3M[3] expected to fund current operating plan into at least 2027

1) Subject to the terms of the definitive agreements between Kiniksa and Regeneron; 50% of ARCALYST Collaboration Operating Profit plus 50% of ARCALYST Licensing Proceeds;
2) Profit Split-Eligible Cost of Goods Sold = total cost of goods sold - amortization of Regeneron milestone payment
3) As used herein the term, "Cash Reserves" means our cash, cash equivalents, and short-term investments (unaudited) as of December 31, 2023
RP = Recurrent Pericarditis, CAPS = Cryopyrin-Associated Periodic Syndromes, DIRA = Deficiency of Interleukin-1 Receptor Antagonist

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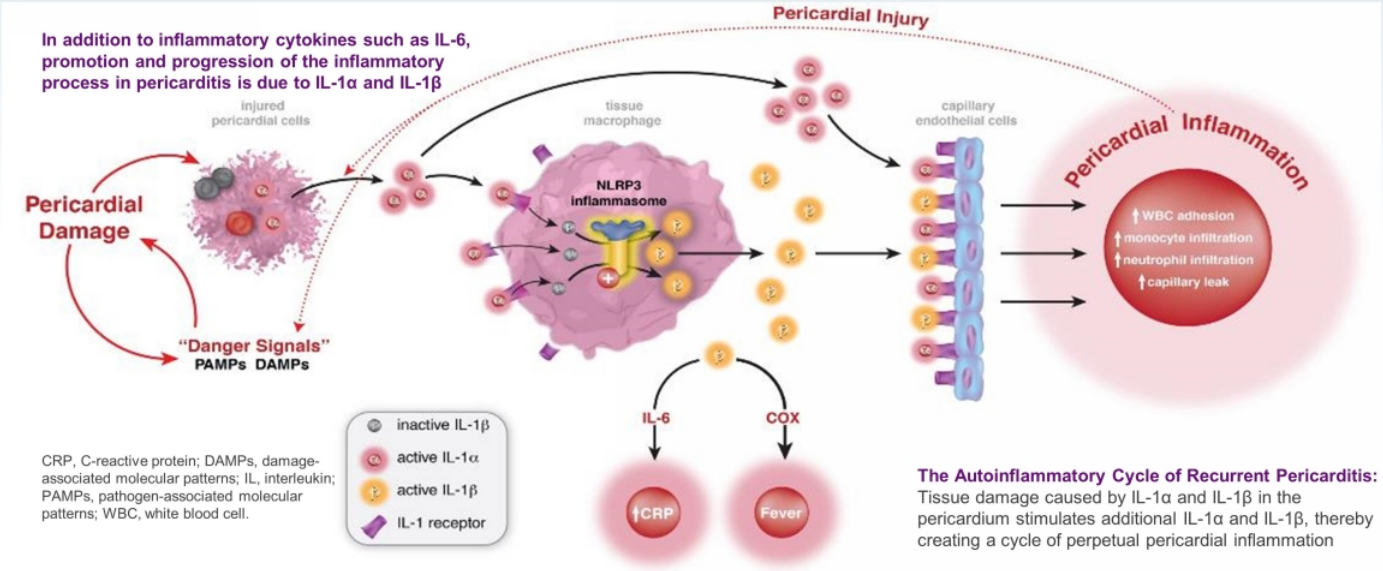
Appendix
ARCALYST (rilonacept)

Role of IL-1α and IL-1β in the Autoinflammatory Cycle of Recurrent Pericarditis



CRP, C-reactive protein; DAMPs, damage-associated molecular patterns; IL, interleukin; PAMPs, pathogen-associated molecular patterns; WBC, white blood cell.

The Autoinflammatory Cycle of Recurrent Pericarditis: Tissue damage caused by IL-1α and IL-1β in the pericardium stimulates additional IL-1α and IL-1β, thereby creating a cycle of perpetual pericardial inflammation

Brucato A, et al. Int Emerg Med 2018 https://doi.org/10.1007/s11739-018-1907-x
Dinarello CA, et al. Nat Rev Drug Discov 2012;11:633-652

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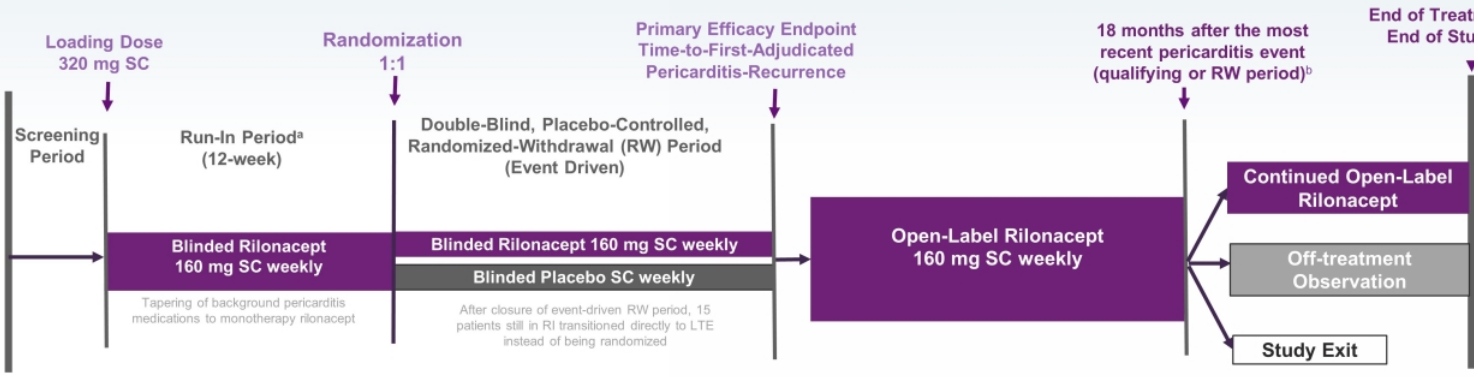
RHAPSODY Design

Event-Driven Pivotal Study

Median rilonacept treatment duration prior to the LTE (RI+RW) was 9 months (range, 3-14)

Long-Term Extension (LTE) (up to 24 months)



[a] The duration of the run-in period was concealed from patients, so that they were blinded to the timing of randomization

[b] For each patient in the LTE, **a decision was made 18 months after the most recent pericarditis recurrence (Qualifying or RW period)** based on clinical status and one of the following actions was taken at the investigator's discretion:

- Continue rilonacept on-study

OR

- Suspend rilonacept treatment and remain on-study for observation (rilonacept rescue for recurrence allowed)

OR

- Discontinue the LTE completely (no further observation)

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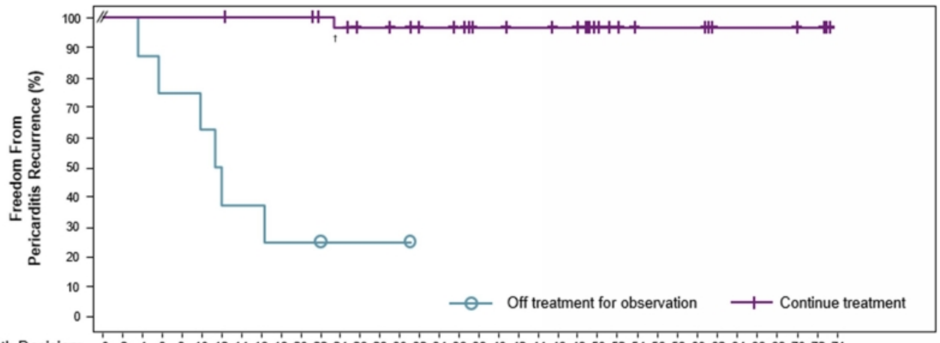
Adapted from: Imazio M, Klein AL, et al. Prolonged Rilonacept Treatment in Rhapsody Long-term Extension Provided Persistent Reduction of Pericarditis Recurrence Risk. Poster 2223 (Presented at AHA Scientific Sessions 2022)

RHAPSODY Long-Term Extension Data Demonstrated Rilonacept Treatmen Beyond 18 months Resulted in Continued Treatment Response[1]



Time (weeks)

Week after 18-Month Decision:	0	2	4	6	8	10	12	14	16	18	20	22	24	26	28	30	32	34	36	38	40	42	44	46	48	50	52	54	56	58	60	62	64	66	68	70	72	74
Continued Rilonacept Treatment, Patients at Risk, n	33	33	33	33	33	33	33	32	32	32	32	30	29	27	27	25	24	23	22	18	18	17	17	16	16	11	9	7	7	7	7	4	4	4	4	4	3	0
Off Treatment for Observation, Patients at Risk, n	8	8	7	6	6	6	4	3	3	2	2	2	1	1	1	1	0																					

†The patient with a recurrence at 23.4 weeks had interrupted rilonacept treatment ~4 weeks prior.

Hazard ratio = 0.02
Log-rank *P*<0.0001
Risk reduction = 98%

	N	Patients with Recurrence,[a] n (%)	Weeks t Recurrenc Median (95
Continued rilonacept treatment	33	1 (3)	NE (NE–NI
Off treatment for observation	8	6 (75)	11.8 (3.7–N

[a]After 18-month decision.
CI, confidence interval; NE, not estimable.

1) Imazio M, Klein AL, et al. Prolonged Rilonacept Treatment in Rhapsody Long-term Extension Provided Persistent Reduction of Pericarditis Recurrence Risk. Poster 2223 (Presented at AHA Scientific Sessions 2022)

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Appendix Out-Licensing Agreements

Out-Licensing Agreements

Partnership with Huadong Medicine Gives Kiniksa Opportunity to Expand Footprint into Asia Pacific Region (Excluding Japan)

- In February 2022, Kiniksa announced a strategic collaboration with Huadong to develop and commercialize ARCALYST and mavrilimumab in Greater China, South Korea, Australia and 18 other countries, excluding Japan
- Kiniksa received a $22M upfront payment and is eligible to receive up to approximately $640M in specified development, regulatory and sales-based milestone along with tiered royalty payments
- Collaboration provided non-dilutive capital, cost-sharing, and additional resources to help accelerate development and commercialization efforts

License Agreement with Roche Genentech for Global Rights to Develop and Commercialize Vixarelimab

- Kiniksa has received $100 million in upfront and near-term payments:
  - $80 million, which was received following the transaction's closing in Q3 2022
  - $20 million, which was received following Kiniksa's last delivery of certain drug supplies to Genentech in Q1 2023
- Kiniksa is eligible to receive up to approximately $600 million in certain clinical, regulatory, and sales-based milestones, before fulfilling upstream financial obligations, of which approximately $585 million remains
- Kiniksa is also eligible to receive royalties on annual net sales ranging from low-double digits to mid-teens, before fulfilling upstream financial obligations
- Proceeds from the transaction to help grow cardiovascular franchise and build autoimmune franchise

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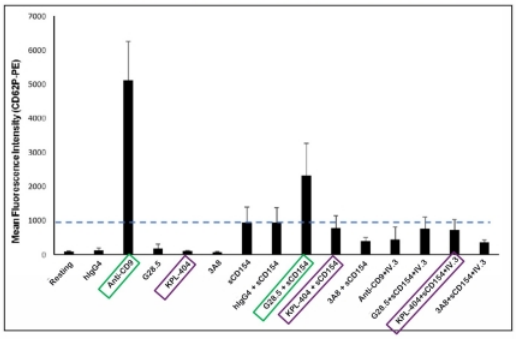
Appendix
Abiprubart

Abiprubart Does Not Cause Platelet Activation or Aggregation *in vitro*

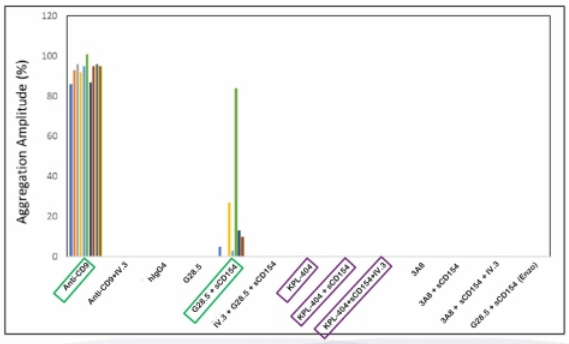
- At least three first-generation IgG1 anti-CD154 mAbs* were associated with thromboembolic events in humans and NHPs[1]
- **Mechanism:** Activation of platelets through cross-linking mediated by IgG-Fc/FcγRIIa interaction
  - Platelet activation observed *in vivo* with anti-CD154 mAbs with active Fc region
  - Platelet activation *in vitro* by anti-CD40 mAbs requires presence of sCD154 and active Fc region
  - Absence of an active Fc-region prevents platelet activation[1,2]

Abiprubart did not cause upregulation of the cell-surface platelet activation marker CD62P
Abiprubart did not induce platelet aggregation in the presence (or absence) of soluble CD154[3]

Abiprubart Alone and in Combination with sCD154 does not increase CD62P Expression on the Platelet Surface



Abiprubart Alone and in Combination with sCD154 does not increase Platelet Aggregation Amplitude (%)

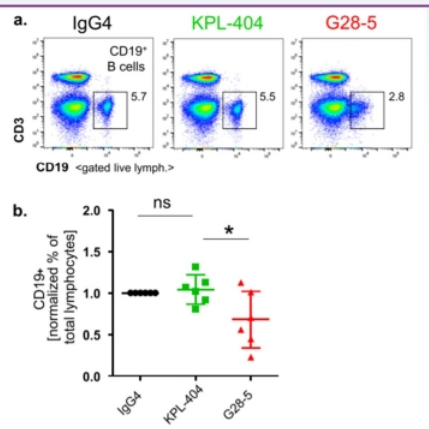


Positive controls:
- G28.5: anti-CD40 mAb – causes sCD40L-dependent platelet activation (Langer et al., Thromb Haemost 2005;93(6):1137-1146)
- Anti-CD9: mAb– causes sCD40L-independent plate activation
- IV.3 - anti-FcγRIIa antibod

*ruplizumab/hu5c8, toralizumab/IDEC-131, ABI793
Sources: 1) Law & Grewal, Advances in Experimental Medicine and Biology, vol 647. Springer; 2) Shock et al., Arthritis Research & Therapy 17, Article Number: 234 (2015); 3) KNSA in-house data

Abiprubart Does Not Reduce B cell Numbers, Activate B Cells, or Induce B Cell Proliferation *in vitro*

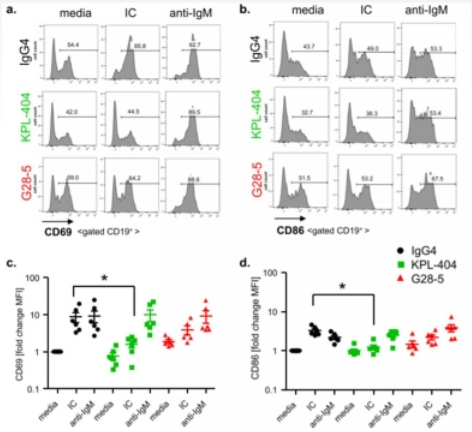
Abiprubart does not reduce B cell numbers in activated PBMCs *in vitro*



G28.5: agonistic aCD40 mAb

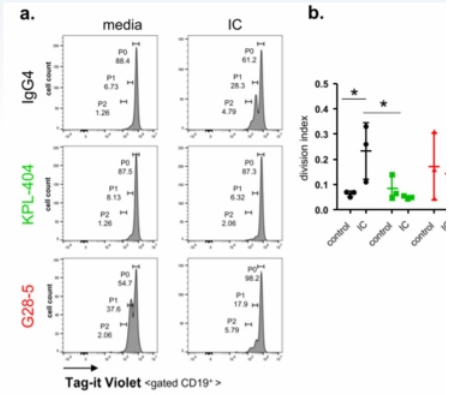
PBMCs were cultured in the presence of 10 µg/ml IgG4 isotype control or anti-CD40 Abs Abiprubart, or the agonistic aCD40 mAb, G28-5 (16–18 h of cell culture)

Abiprubart does not induce B cell activation



PBMCs were cultured in the presence of 10 µg/ml IgG4 isotype control or anti-CD40 Abs Abiprubart, or G28-5 (16–18 h of cell culture). Cells were left unstimulated (media control) or stimulated with CD3/CD28 cross-linker IC or F(ab')2 goat anti-human IgM (anti-IgM)

Abiprubart does not induce B cell proliferation *in vitro*



PBMCs were labeled with a cell proliferation tracker dye (Tag it Violet) and cultured for 5 days in the presence of 10 µg/ml IgG4 isotype control Ab or anti-CD40 Abs—Abiprubart and G28-5. Cells were left untreated (media control) or stimulated with anti-CD3/CD28 cross-linking reagent ImmunoCult (IC)

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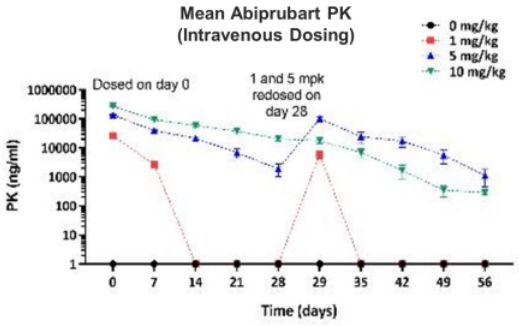
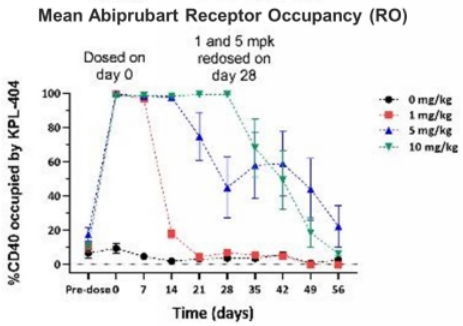
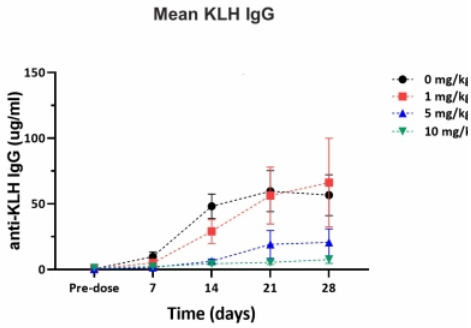
*Marken et al, Arthritis Res Ther. 2021 Jan 21;23(1):36. doi: 10.1186/s13075-021-02425-x.

Abiprubart Demonstrated Prolonged Suppression of TDAR Response in a Non-Human Primate Model





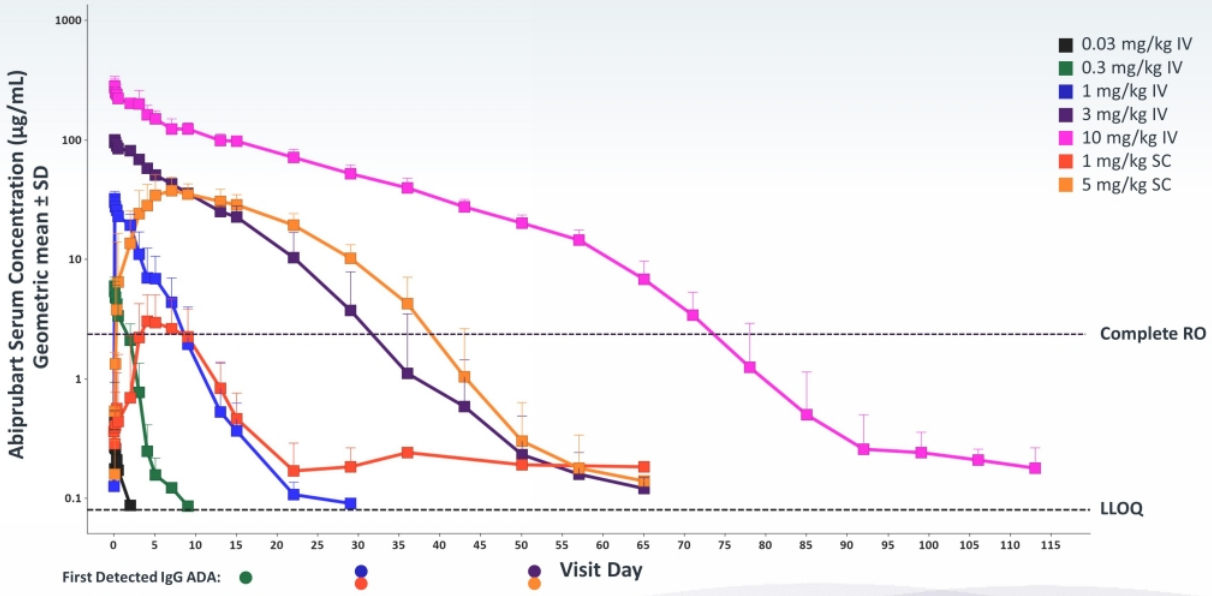


Showed linear pharmacokinetic profile with low variability between non-human primate subjects (n=7)	*Abiprubart achieved 100% receptor occupancy for 2 weeks in all animals at 5mg/kg and 4 weeks in all animals at 10mg/kg*	*Complete suppression of primary T-cell depende antigen response correlated with 100% recepto occupancy*

Source: Muralidharan et al. Preclinical immunopharmacologic assessment of KPL-404, a novel, humanized, non-depleting antagonistic anti-CD40 monoclonal antibody. J Pharmacol Exp Ther. 2022, 381(1):12-21
TDAR = T-cell dependent antibody response; KLH = keyhole limpet hemocyanin

Final Data from Abiprubart Single-Ascending-Dose Phase 1 Study

Pharmacokinetic profiles for abiprubart

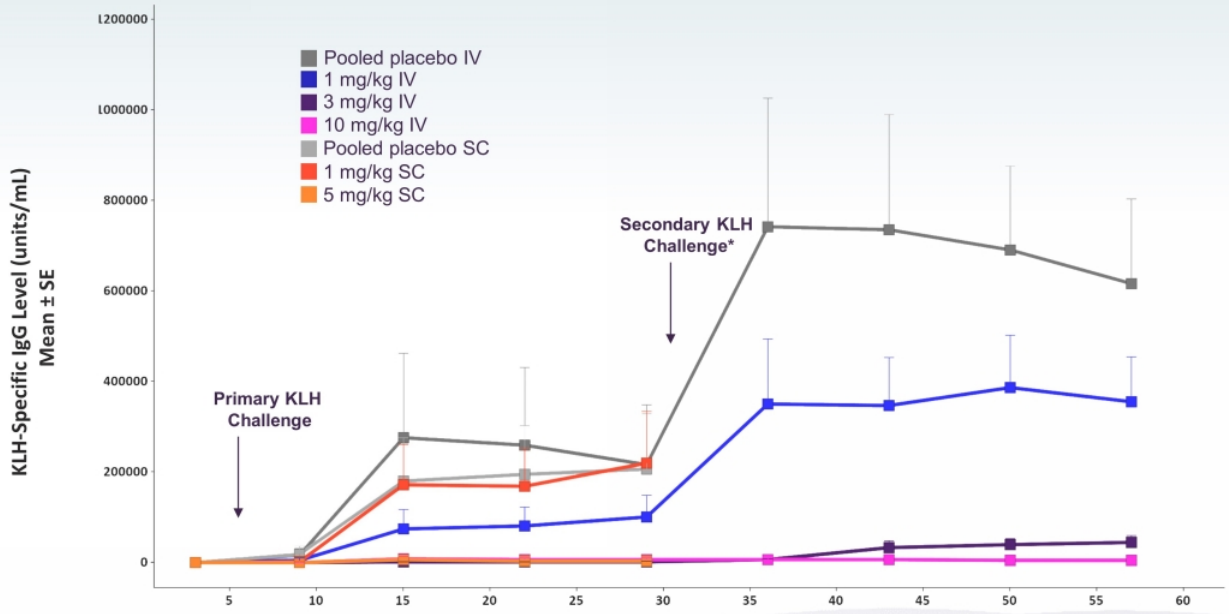


Source: Samant M, Ziemniak J, Paolini JF. First-in-Human Phase 1 Randomized Trial with the Anti-CD40 Monoclonal Antibody KPL-404: Safety, Tolerability, Receptor Occupancy, and Suppression of T-Cell-Dependent Antibody Response. J Pharmacol Exp Ther. 2023 Dec;387(3):306-314.

SD = standard deviation (upward bars depicted); IV = intravenous; SC = subcutaneous; LLOQ = lower limit of quantitation; ADA = anti-drug antibody

Final Data from Abiprubart Single-Ascending-Dose Phase 1 Study

T-Cell Dependent Antibody Response (TDAR) for KLH antigen challenge



*Only IV cohorts were rechallenged with KLH on day 29

Source: Samant M, Ziemniak J, Paolini JF. First-in-Human Phase 1 Randomized Trial with the Anti-CD40 Monoclonal Antibody KPL-404: Safety, Tolerability, Receptor Occupancy, and Suppression of T-Cell-Dependent Antibody Response. J Pharmacol Exp Ther. 2023 Dec;387(3):306-314.

KLH = keyhole limpet hemocyanin

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Corporate Presentation

JANUARY 2024